

## Primary Mucinous Carcinoma Scalp: A Rare Case Report With Review of Literature

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### ABSTRACT

Primary mucinous adenocarcinoma of scalp is a rare malignant neoplasm with predilection to head and neck area. This tumor mostly occurs in sixth or seventh decade with tendency of local recurrence. The distant metastasis is rarely seen. It is essential to differentiate the primary neoplasms from metastatic neoplasm arising from breast, gastrointestinal tract and other organs because the prognosis and management differs drastically. We present a case report of a 43-year-old female with swelling in scalp diagnosed as primary mucinous carcinoma, without any systemic dissemination. We review the literature about primary and metastatic mucinous neoplasms in order to better understand, identify and manage this entity.

**Keywords:** Primary mucinous adenocarcinoma, Metastatic neoplasm, Recurrence, Dissemination, Prognosis.

### 1. Introduction

Primary cutaneous mucinous carcinoma (PCMC) is a rare mucin-producing malignancy derived from epithelial glandular structures, first described in 1952 by Lennox et al, followed by Breiting et al, Levy et al, Mendoza & Helwig [1]-[4]. The tumor most often presents in the sixth or seventh decade with a predilection for the head and neck [5]-[7]. It tends to locally recur, but rarely metastasizes [1],[2],[5]. On occasion, it affects other sites including scalp, face, ear, axilla, thorax abdomen, groin, foot, hand and vulva [8]. It is usually solitary and grows slowly over several months or even years [9]. The main challenge is to differentiate primary neoplasm from metastatic mucinous adenocarcinoma originating from other primary sites like breast, gastrointestinal tract is a diagnostic dilemma.

### 2. Case Report

A 43-year-old female patient came with history of hair fall 11 years back. She took injection of unknown substance from a quack. Later, she had dengue followed by an episode of brain hemorrhage and went in coma for one and half months. She gradually recovered and now presented with history of scalp swelling which was gradually increasing from 6 months. There was no lymphadenopathy or features of systemic dissemination. Fine needle aspiration cytology showed loose clusters of basaloid cells in a background of proteinaceous fluid. Possibility of skin adnexal tumor cystic eccrine hidradenoma was suggested.

We also advised excision biopsy of the same. The excised specimen was (2x1.5x1) cm and was gray, brown solid with glistening areas on cut section. On microscopic examination nests of tumor cells were seen in the pools of mucin and this mucin was separated by thin fibrous septa. Individual tumor cells were small to medium sized, hyperchromatic with scant cytoplasm. These cells were relatively uniform in size and shape, showing mild atypia with no evidence of mitosis. The features were suggestive of mucinous carcinoma. As patient had not undergone any previous investigations for the swelling, we advised her to undergo mammography and computerized

tomography abdomen and pelvis to rule out metastasis of carcinoma breast and gastro-intestinal neoplasm. The mammography of bilateral breast was normal, and computerized tomography scan showed simple cortical cyst measuring (1.8x1.8) cm arising from the upper pole cortex, right kidney and bulky uterus showing features of pelvic inflammatory disease. Rest findings were unremarkable.

### 3. Discussion

Primary cutaneous mucinous carcinoma is a tumor seen more frequently in men between ages of 50-70 years, as a slow-growing, painless, nodular, red/gray/purple lesion that may be ulcerated/crusted or with features of telangiectasia [9]. Histologically, the tumor is situated in the dermis and often involves the subcutaneous fat. Lakes of pale-staining mucin with suspended islands of tumor cells compartmentalized by delicate fibrous septa are seen. The mucin is Periodic acid-Schiff (PAS) positive diastase resistant, hyaluronidase-resistant. Alcian blue positive (pH 2.5) and stains with Mucicarmine and colloidal iron [8].

The primary challenge in diagnosis lies in differentiating these rare primary skin neoplasms of sweat gland origin from the more frequent mucinous secondary deposits to the skin from primaries elsewhere. Mucin-producing primary tumors are known to originate in the breast, gastrointestinal tract, lung, kidney, ovaries, pancreas and prostate [10],[11]. Metastatic lesions from the breast and colon are most likely to mimic mucinous carcinoma of the skin, knowing the fact that 19% of the men with colon cancer and 6% of the women with breast cancer have metastatic skin disease [12],[13].

To differentiate secondary deposits, from these two sites from primary mucinous skin carcinoma just on basis of morphological evaluation alone is almost impossible. Therefore, the differentiation is mainly based on ruling out the presence of another primary malignant site through a full oncological evaluation, and histo-pathological characteristics of the mucinous lesion. Histologic distinction between primary and secondary tumors may be impossible, albeit the latter may show certain features supporting metastasis such as less abundance of mucin, large tumor clusters and sheets with predominance of malignant epithelial cells over mucin, and lack of honey combing due to absence of fibrous septae in between the mucinous lakes [14]. Immunohistochemistry positive for CK7 & negative for CK-20 rules out metastasis from stomach and colon [15]. Estrogen receptors & Progesterone receptors both are positive in breast carcinoma and primary cutaneous mucinous carcinoma but proper clinical and radiological examination along with no evidence of any lesion in breast rules out metastatic breast carcinoma.

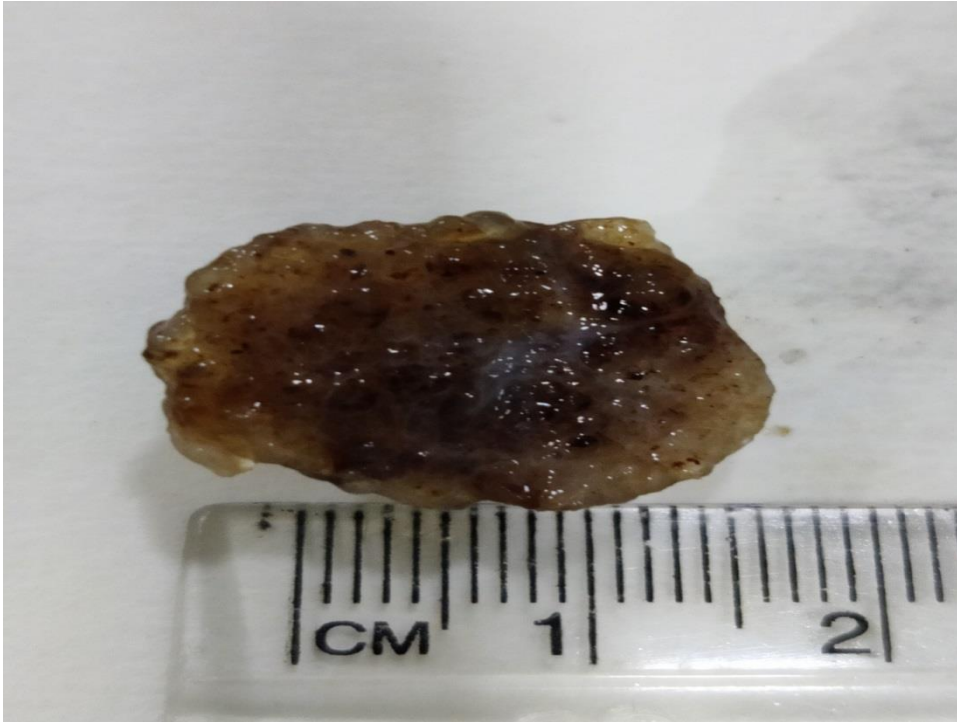
As for treatment, surgical excision of primary eccrine mucinous adenocarcinoma of the skin is the therapeutic mainstay in most cases. Because of the recurring nature of the tumor, adequate excision with wide margins (at least 1cm) is advocated. Moh's micrographic surgery can be a particular advantageous treatment modality in this setting. These tumors are generally resistant to radiotherapy and chemotherapy [10],[12] So, these patients should be counseled for frequent follow up because of recurrence, regional lymphadenopathy and to rule out metastasis.

### 4. Conclusion

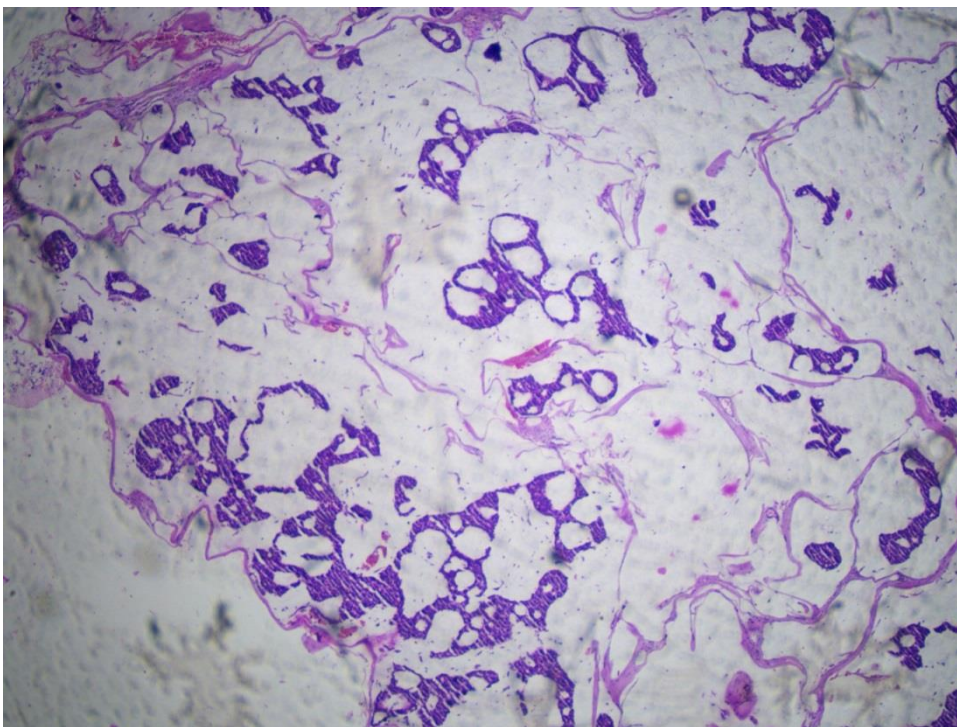
Before diagnosing a primary cutaneous mucinous adenocarcinoma, one should rule out metastasis from breast, gastrointestinal tract and other organs. They have low grade malignant potential with recurrences and limited to

regional lymph node metastasis only. Distant metastasis is rarely seen. As it is difficult to differentiate primary from metastatic adenocarcinoma, a meticulous approach with patient's proper counselling, follow up and clinicopathological correlation for proper treatment and management of this entity is must.

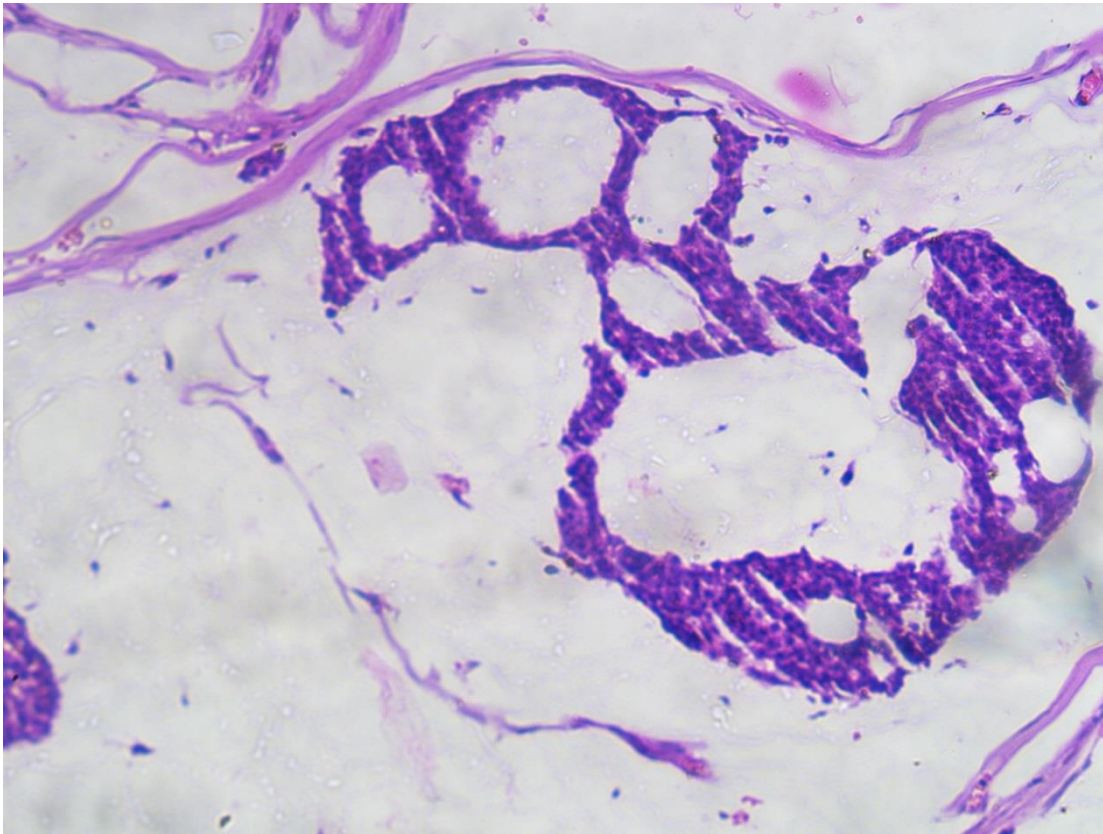
#### **Legends Mucinous Carcinoma Scalp**



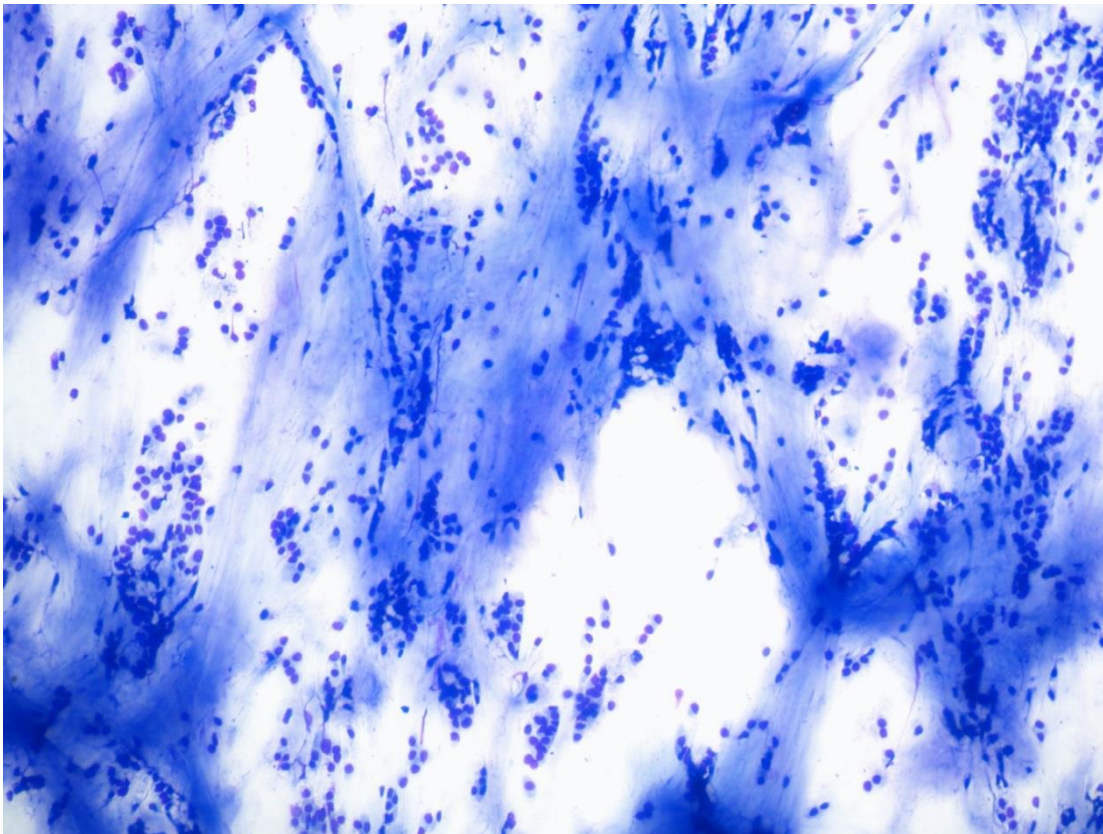
**Fig.1.** Gross specimen of mucinous carcinoma scalp showing gray, brown glistening areas on cut section



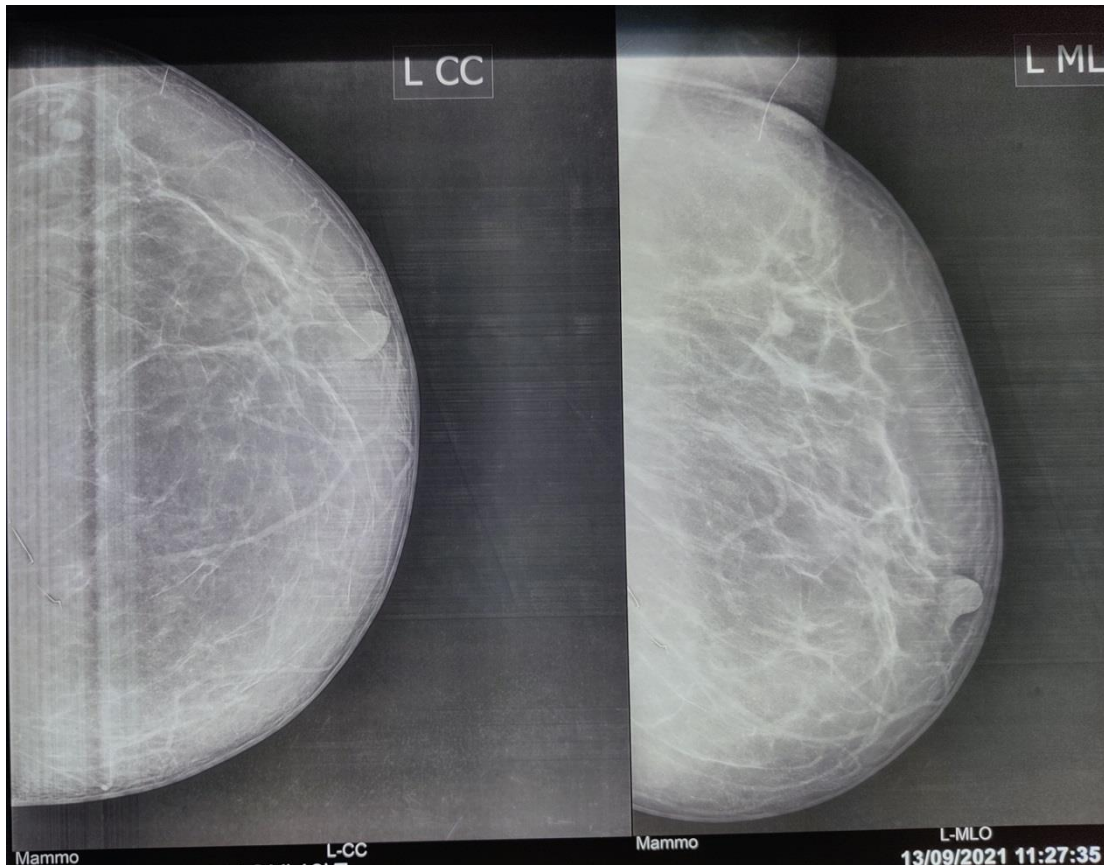
**Fig.2(A).** Histology 10x10 (H&E) Mucinous carcinoma scalp swelling showing lakes of mucin separated by delicate fibrous septa and nests, ducts and cribriform pattern of tumor cells



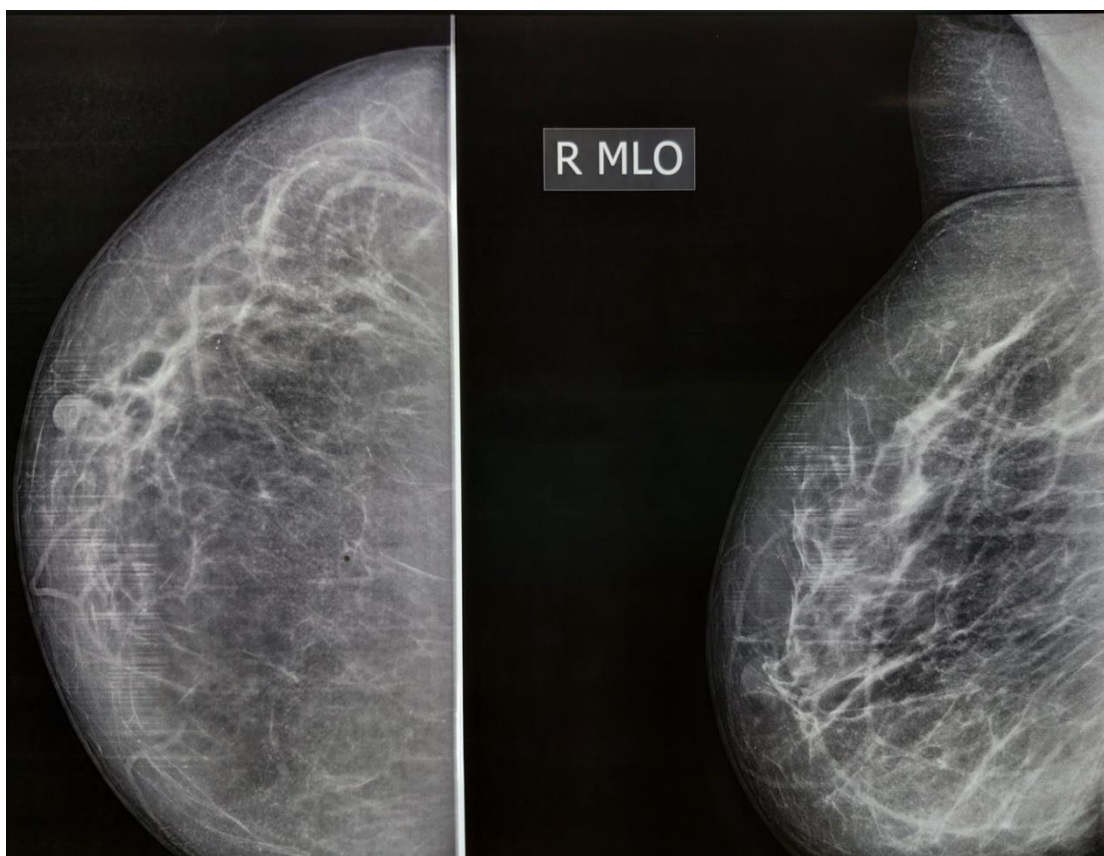
**Fig.2(B).** Histology 40x10 (H&E) Tumor showing neoplastic cell proliferation in cribriform pattern



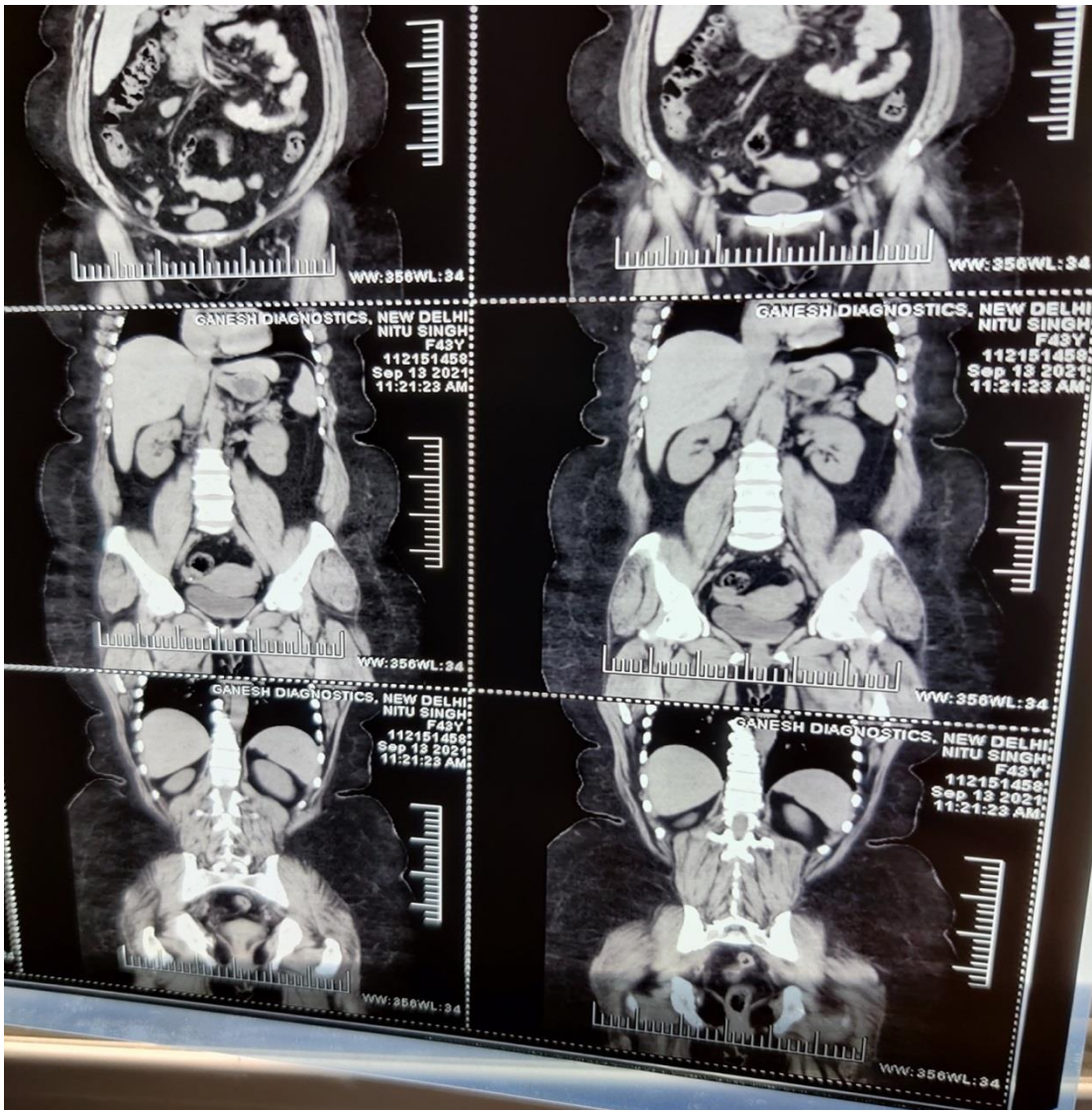
**Fig.3.** Cytology mucinous carcinoma 10x10 (Giemsa) Cellular smears showing basaloid tumor cells within dispersed pools of mucin



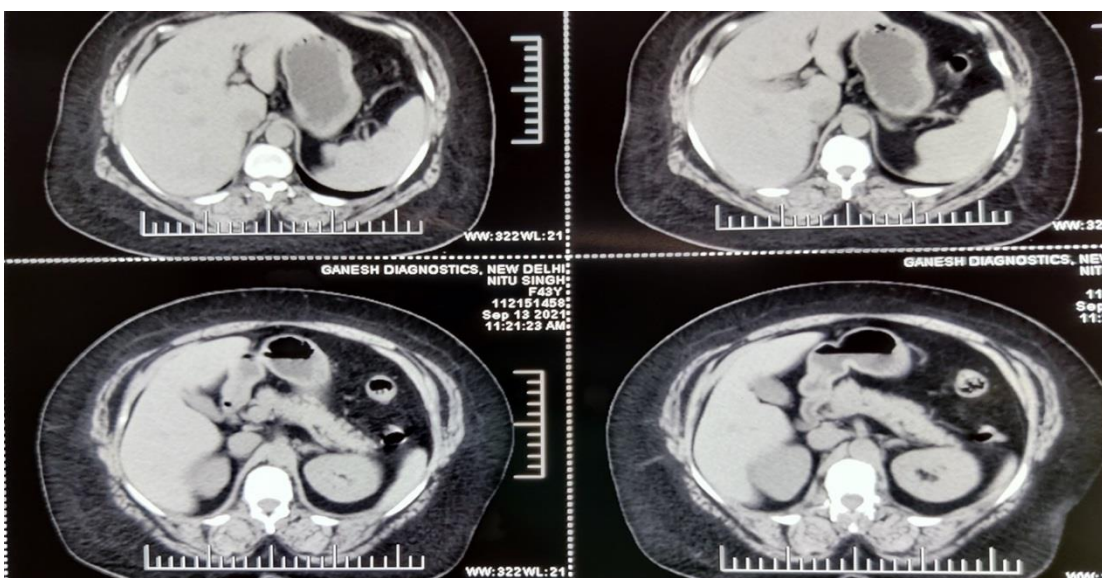
**Fig.4(A).** Mammography left breast, craniocaudal view showing fatty fibro glandular pattern



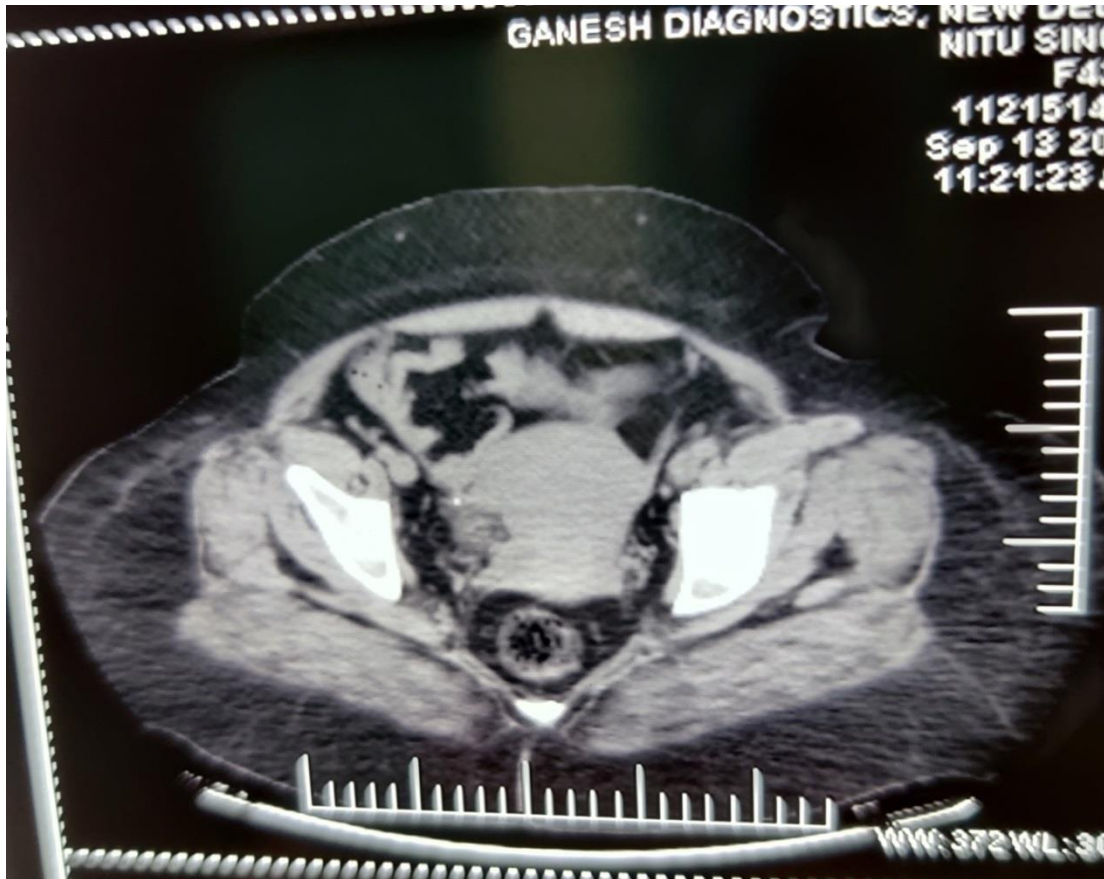
**Fig.4(B).** Mammography right breast, mediolateral oblique view showing normal fibro glandular parenchyma



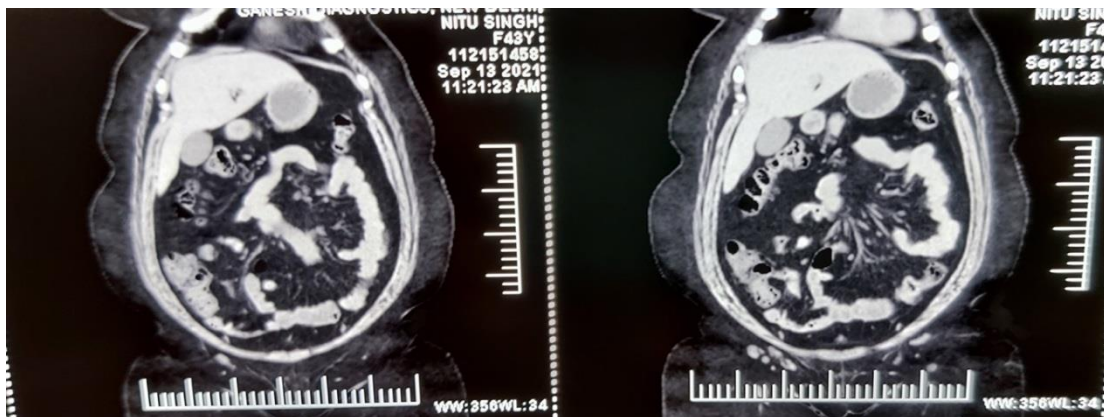
**Fig.5(A).** CT abdomen and pelvis (plain) showing simple cortical cyst measuring 1.8 x1.8 cm arising from the upper pole cortex right kidney



**Fig.5(B).** CT abdomen and pelvis (plain) showing normal liver, pancreas and gall bladder



**Fig.5(C).** CT abdomen and pelvis (plain) showing bulky uterus 8.8 (CC) X 6.9 (RL) X 5.8 (AP) cm. with features of pelvic inflammatory disease



**Fig.5(D).** CT abdomen and pelvis (plain) showing normal appendix and large bowel loops distended with fecal matter. No free fluid is seen in the peritoneum

## Declarations

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### Competing Interests Statement

*The authors declare no competing financial, professional and personal interests.*

### **Consent for publication**

*Authors declare that they consented for the publication of this research work.*

### **Declaration of Patient Consent**

*The authors certify that they have obtained all appropriate consent forms. In the form the patient(s) has/have given his/her consent for his/her images and other clinical information to be reported in the journal. The patients understand that their name and initials will not be published, and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.*

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